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Case Study

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CASE REPORT ON: TOLOSA HUNT SYNDROME

Maheshwari V.[1*], Syam Prakash K. R.[1*], Shahina K. P.[2*] and Manju Mohan[3*]

[1]PHARM D Interns, Department of Pharmacy Practice, Nandha College of Pharmacy, Erode, Tamil Nadu, India.
[2]Clinical Pharmacist, Department of Clinical Pharmacy, IQRAA International Hospital and Research Centre, Calicut, India.
[3]Pharmacist, Department of Pharmacology, IQRAA International Hospital and Research Centre, Calicut, India.

***Corresponding Author: Maheshwari V.**

PHARM D Interns, Department of Pharmacy Practice, Nandha College of Pharmacy, Erode, Tamil Nadu, India.

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ABSTRACT

Tolosa-Hunt Syndrome(TSH) is a rare disease with a limited number of cases reported in the literature. It is caused by non-specific inflammation of the cavernous sinus, superior orbital fissure and the apex of the orbit. It typically presents with orbital pain associated with palsy of the third, fourth, and or sixth cranial nerve and quick response to steroid treatment. We present an interesting case of Tolosa-Hunt syndrome in a 67 year old male patient who was admitted with periorbital pain, severe headache, diplopia and right L R palsy and responded well to high-dose steroids and in a few days had significant improvement in his retro-orbital pain, and diplopia.

KEY WORDS: Tolosa- Hunt syndrome, periorbital pain, plasy, diplopia.

INTRODUCTION

Tolosa Hunt syndrome (THS) is described as severe and unilateral periorbital headache associated with painful and restricted eye movements. Synonyms for Tolosa Hunt syndrome include painful ophthalmoplegia, recurrent ophthalmoplegia, ophthalmoplegia syndrome. Tolosa-Hunt syndrome (THS) is a rare steroid-responsive etiology for painful ophthalmoplegia. And it is usually idiopathic and is thought to be from non-specific inflammation in the region of the cavernous sinus and/or superior orbital fissure. However, traumatic injury, tumors, or an aneurysm could be the potential triggers.

Severe orbital or peri-orbital pain, constant or episodic, is the initial feature of THS that usually resolve spontaneously or under treatment but tend to relapse. Diplopia related to ophthalmoparesis or disordered eye movement occurs when the oculomotor (III), trochlear (IV) and abducens (VI) nerves are damaged by granulomatous inflammation. Signs of incomplete nerve palsy with or without pupillary sparing may be present. A ptosis can be observed due to oculomotor paralysis.[1]

Pathophysiology

The Tolosa-Hunt syndrome is caused by an inflammatory process of unknown etiology. On histopathology, there is a nonspecific inflammation of the septa and wall of the cavernous sinus, with a lymphocyte and plasma cell infiltration, giant cell granulomas, and proliferation of fibroblasts.[2,3] The inflammation produces pressure and secondary dysfunction of the structures within the cavernous sinus, including cranial nerves III, IV, and VI, as well as the superior divisions of cranial nerve V.

International headache society (IHS) diagnostic criteria

Tolosa Hunt Syndrome also finds a place in the IHS Classification ICHD-3 Beta, in part three, under Painful cranial neuropathies and other facial pains. The IHS lays down diagnostic criteria for THS which have high sensitivity (approximately 95% to 100%) but low specificity (approximately 50%).[4] They are summarized as follows:

- Unilateral headache
- Includes both of the following:

1. Presence of granulomatous inflammation of the cavernous sinus, superior orbital fissure or orbit, as seen on MRI or biopsy
2. Palsies of one or more of the oculomotor nerves (cranial nerves III, IV, and/or VI) on the same side

- Corroboration of the cause as evidenced by both of the following:

1. Palsies of cranial nerves III, IV, and/or VI have followed headache in two weeks or less, or have developed simultaneously with a headache
2. Localization of a headache around the eye on the same side

- Not better explained by any other headache etiology

Treatment

Glucocorticoids have long been the recommended treatment for Tolosa Hunt Syndrome. Little consideration has been given to alternative therapies, probably due to the typical rapid response of glucocorticoids (pain usually resolves within 24 to 72 hours after starting treatment).

CASE REPORT

A 67 years old male patient with history of DM, HTN on treatment and admitted with periorbital pain and insomnia, pain was sharp and continuous, patient have severe headache and diplopia (seeing two images of an object, double vision), and history of herpes neuralgia.

On general examination patient was conscious and oriented, BP was 140/90 mmHg, pulse rate was 78 beats/min and patient have right L R plasy which is associated with dysfunction of cranial nerve V1. On laboratory investigation patient RBS was high 200mg/dL and HBA1C level was 11.3% which indicate patient was poorly diabetic. On CSF analysis sugar was slightly increased 121mg/dL and meningal infection was ruled out by CSF study. We performed the following complementary studies: complete blood count showed (white cell count of 7, 200cells/cumm) and haemoglobin of 13.2 gm/dL (haematocrit 50%). Erythrocyte sedimentation rate was 18mm/hr. Serum chemistry including liver function tests was normal. Vasculitis markers, thyroid function tests, anti-thyroglobulin, anti-microsomal receptor were normal.

MRI Brain: Shows old lacunar infracts in right cerebellar hemisphere, right lentiform nucleus. Age related degenerative changes in brain parenchyma.

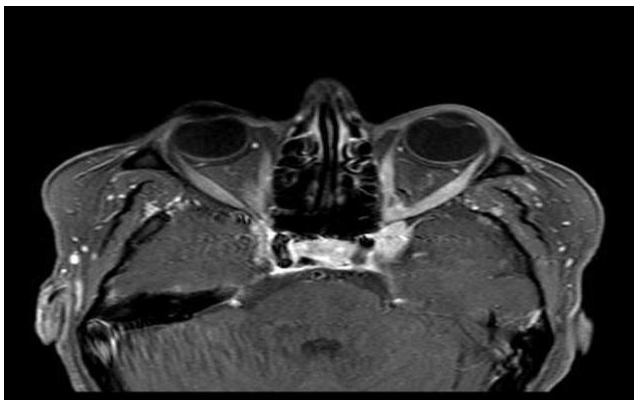

Fig 1: MRI Brain scan.

MRI brain showed old lacunar infracts only and MRI BRAIN CONTRAST showed homogenously enhancing soft tissue in anterior part of right cavernous sinus and adjacent part of superior orbital fissure- likely Tolosa Hunt Syndrome. Patient was treated with IV METHYL PREDNISOLONE 500mg in 250mL NS for 3 days following oral steroid T. PREDNISOLONE 40mg OD and diabetic control by insulin therapy. Patient well responded to steroids and in a few days had significant improvement in his retro-orbital pain and diplopia. Patient was symptomatically better and discharged.

DISCUSSION

Tolosa Hunt syndrome is one of the rare disorder, recognized by the National Organisation for Rare Disorders (NORD). It is also included as one of the painful cranial neuropathies by the International Headache Society (IHS) in its headache classification. Tolosa Hunt syndrome was first described in the year 1954 by Dr. Eduardo Tolosa, a Spanish neurosurgeon.[1] in a patient with unilateral recurrent painful ophthalmoplegia involving cranial nerves III, IV, VI and V1. In our case patient was Right L R palsy which is associated with dysfunction of cranial nerve V1.

Tolosa-Hunt syndrome is characterized by severe periorbital headaches, along with decreased and painful eye movements (ophthalmoplegia). Symptoms usually affect only one eye (unilateral). In most cases, affected individuals experience intense sharp pain and decreased eye movements. Symptoms often will subside without intervention (spontaneous remission) and may recur without a distinct pattern (randomly). Affected individuals may exhibit signs of paralysis (palsy) of certain cranial nerves such as drooping of the upper eyelid (ptosis), double vision (diplopia), large pupil, and facial numbness. The affected eye often abnormally protrudes (proptosis).

Tolosa Hunt syndrome follows a variable course that can last from days to weeks to months. Recurrences are common and can either be unilateral or bilateral. Other causes must be excluded by appropriate investigations. The retro-orbital pain has been shown to completely resolve within 72 hours of onset of steroid treatment, but the time needed for normalization of the cranial nerve palsies has been broad with an average of 26 days.

Neuro imaging and LCR are important in differential diagnosis. It is also important to analyse complete blood count, ESR protein, C-reactive protein, complete biochemical tests, including functional tests of the thyroid and liver. Chest X-ray and sinus CT are sometimes indicated. Brain and orbital imaging includes MRI and CT. MRI (T2- weighted, including fat suppression, STIR sequence) helps to identify high signals in an extra-ocular active muscle. Inflammatory causes of painful ophthalmoplegia include those due to a specific infectious agent. It is essential to conduct a careful examination of CSF and obtain cultures (bacterial, fungal, mycobacterial).[5]

High-dose corticosteroid therapy is the first-line therapy for Tolosa Hunt syndrome given its inflammatory pathology.[1] High-dose corticosteroid therapy is the first-line therapy for Tolosa Hunt syndrome given its inflammatory pathology.[1] There is no evidence sufficient for the appropriate dose, route of administration and duration of therapy.[6] A remarkable

feature of glucocorticoid therapy is the rapid resolution of orbital pain within 1-3 days, which also serves as confirmation of a diagnosis.[7] Our patient also has same way of treatment, IV METHYL PREDNISOLONE 500mg in 250ml NS for 3 days following oral steroid T. PREDNISOLONE 40mg OD. After an initial high-dose of corticosteroid, an oral taper over the course of several weeks is recommended, along with regular follow-up with subsequent MRI studies to document the resolution of the disease. Immunosuppressive drugs are the other therapeutic modality of Tolosa-Hunt syndrome. The recurrence of the disease occurs in about 50% of patients, which is quite alarming.[8]

CONCLUSION

Tolosa-Hunt syndrome (TSH) is a rare disease, its etiopathogenesis is unknown, it is manifested clinically by unilateral orbital pain associated with simple or multiple oculo motor paralyses, which resolves spontaneously but may recur.

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